Multilocular cystic nephroma is an unusual entity of uncertain etiology that can affect children and adults equally. This lesion is believed to be part of a spectrum of renal cystic disorders with multilocular cyst and cystic Wilms’ tumor representing the two extremes and usually presents with a benign character. We report a 22-year-old female who presented with a painful right renal mass and gross hematuria. After a series of examinations including abdominal ultrasound, computed tomography, and angiography, she underwent partial nephrectomy under the impression of benign lesion. The pathologic examination showed that it was a multilocular cystic nephroma. At the 11-month follow-up, the patient was completely asymptomatic and free of recurrence and metastasis.

Key Words: multilocular cystic nephroma, partial nephrectomy

Multilocular cystic nephroma is a very rare benign cystic tumor of the kidney that is almost uniformly unilateral and compresses the adjacent normal renal tissue [1]. Several names have been used to describe this kidney lesion, including benign multilocular cyst, multilocular renal cyst, and cystic nephroma. Patients usually present with an abdominal mass with or without abdominal pain and hematuria [2]. We report a case of unilateral multilocular cystic nephroma that was successfully treated with partial nephrectomy.

**CASE PRESENTATION**

A 22-year-old female presented with a painful right renal mass and gross hematuria. Biochemical parameters and renal function were within normal ranges. Urinalysis showed obvious hematuria and urine cytology was negative for malignancy. Ultrasound showed a large complex cystic mass occupying the right kidney from the mid to the lower portion. Computed tomography (CT) showed a 9.1 × 6.3 cm complex cystic mass in the right kidney (Figure 1). Arterial angiography showed hypovascularity of the mass (Figure 2). Abdominal exploration was carried out through a subcostal approach. Partial nephrectomy was performed and the remaining portion of the right kidney was spared. On cut section, the mass comprised well-differentiated areas of cysts filled with serous fluid and occasionally appeared hemorrhagic (Figure 3). The cysts did not communicate with each other or with the collecting system. Pathologic examination showed multilocular cysts lined by flattened to cuboidal epithelium, and they were separated by cellular spindle cell stroma (Figure 4). No solid area or necrosis was present. At the 11-month follow-up, the patient was completely asymptomatic with normal renal function and free of recurrence.
DISCUSSION

Multilocular cystic nephroma can occur in children and adults. The lesions typically have a bimodal distribution. Most pediatric cases are seen in girls older than 4 years or in boys younger than 4 years. Multilocular cystic nephromas occur with a female predominance in adults [3]. The peak incidence in adults occurs during the second decade with another peak in the fifth and sixth decades of life. The common symptoms and signs include abdominal mass with or without abdominal pain, hematuria, and hypertension.

In 1956, Boggs and Kimmelstiel established the criteria for the diagnosis of a multilocular cyst [4]. The criteria include: (a) a multilocular mass; (b) no communication between cysts; (c) cysts lined by epithelium; (d) no communication between cysts and pelvis; (e) remaining kidney essentially normal; and (f) no normal nephrons in the septa of cysts. In 1989, Joshi and Beckwith further reformulated the criteria by specifying that the: (a) tumor is composed entirely of cysts and their septa; (b) cystic nephroma is a discrete...
well-demarcated mass; (c) septa are the sole solid component and conform to the outlines of the cyst without expansive nodules; (d) cysts are lined by flattened, cuboidal, or hobnail epithelium; and (e) septa contain fibrous tissue in which well-differentiated tubules may be present [5].

Differential diagnoses range from polycystic kidney, hydronephrotic kidney, nephroblastomas, Wilms’ tumor, mesoblastic nephroma to cystic renal cell carcinoma (RCC). Ultrasound, dynamic CT, and dynamic magnetic resonance imaging cannot reliably distinguish between malignant and benign cystic tumors. Renal angiography is the most reliable imaging modality for distinguishing cystic RCC from benign cystic lesions by the presence or absence of tumor vessels. However, the diagnosis may not be confirmed even if intraoperative specimens are obtained for frozen section examination.

As preoperative imaging and intraoperative frozen-section analysis cannot distinguish cystic nephroma from malignant cystic RCC, surgical intervention is the only effective method to differentiate cystic nephroma from a malignant lesion of the kidney. Nephrectomy is an adequate treatment, with no need for chemotherapy and radiotherapy [6]. Thus, a definitive diagnosis can usually be made from the pathology results when the operation is finished. Nephron sparing technique is another appropriate choice for these patients if complete excision is possible [7]. However, based on our experience with this case, we suggest that renal-sparing surgery be considered first if preoperative radiologic study strongly supports the appearance of a benign tumor and is technically feasible.

**REFERENCES**

罕見的多腔室囊性腎組織腫瘤接受腎臟保留手術治療 — 病例報告

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一位二十二歲女性無任何慢性疾病病史，至本院求診之主訴為右側腰部發現一疼痛腫塊及出現明顯血尿，腹部超音波，電腦斷層及血管攝影顯示一低血流性多囊性腎組織腫瘤，病患未採腎全切除術，而採用腎臟保留手術治療，病理報告為多腔室囊性腎組織腫瘤，病患目前仍於本院追蹤且狀況良好並無腫瘤復發現象。

關鍵詞：多腔室囊性腎組織腫瘤，腎臟保留手術
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